Bilateral Persistent Pharyngo-Stapedial Arteries Revealed during Evaluation of a Carotid-Cavernous Fistula

3 Tesla MR Angiographic Aspects

J. JEHL, L. JEUNET*, M. BERRAIAH, J.-F. BONNEVILLE

*Department of Neuroradiology and *Department of Orthopaedic surgery, University Hospital Jean Minjoz, Besançon, France*

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Summary

A 33-year-old woman was evaluated for a right carotid-cavernous fistula revealed by a proptosis and chemosis of the right eye. The initial angiogram showed a left persistent pharyngo-stapedial artery (Ph-SA). A temporal bone CT suggested bilateral pharyngo-stapedial artery persistence. The right Ph-SA was not opacified in the first angiogram because of the high degree of shunting in the fistula. Four months later the patient was admitted for treatment of the carotid-cavernous fistula. In the meantime, the fistula had altered, with spontaneous thrombosis of the ophthalmic vein, and decrease of the vascular steal, explaining that the right Ph-SA was clearly visible on the angiogram performed during the procedure.

The carotid-cavernous fistula was completely occluded with five detachable coils. The follow-up included 3 Tesla MR angiography that showed complete closure of the fistula with preservation of the right ICA and bilateral persistent pharyngo-stapedial arteries.

Introduction

The stapedial artery is a transient vessel, present during normal fetal development, but that regresses in the 24 mm embryo [1]. Post embryonic persistence of the stapedial artery is rare and usually unilateral. Most patients are asymptomatic but symptoms of tinnitus or hearing loss are possible. A persistent stapedial artery (PSA) may also complicate middle ear surgery [2,3]. The pharyngo-stapedial artery is a rare variant of stapedial artery persistence.

Case Report

A 33-year-old caucasian woman was referred to the neuroradiologist for lower limbs angiography. The patient had a history of defenestration with polytraumatism two years earlier. The angiography was needed before rebuilding surgery of her left foot and turned out to be normal. Moreover, clinical examination by the neuroradiologist revealed a proptosis of the right eye with chemosis.

Carotid angiography was performed and showed a right high-flow carotid-cavernous fistula, that drained into the inferior petrosal sinus, superior ophthalmic vein, Brechet sinus and cortical veins. The left carotid angiography incidentally revealed an anomalous vessel arising from the cervical part of the internal carotid artery and running superiorly through the middle ear to supply the middle meningeal artery. A diagnosis of left persistent pharyngo-stapedial artery was made, associated with an unusual emergence of the ascending pharyngeal artery from the internal carotid artery. A temporal bone CT showed the probable bilaterality of the anomaly, with bilateral absence of

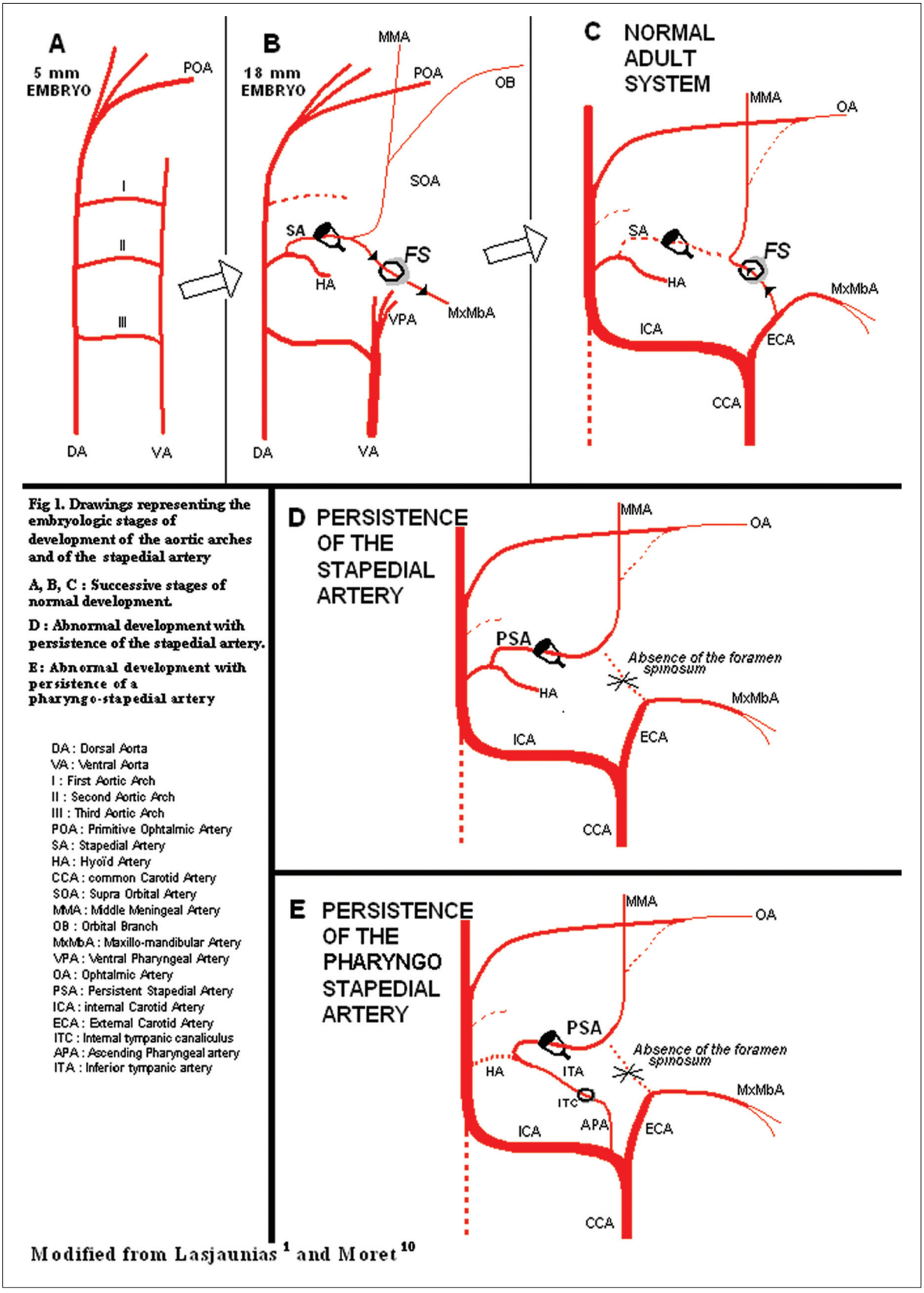

Fig 1. Drawings representing the embryologic stages of development of the aortic arches and of the stapedial artery

A, B, C : Successive stages of normal development.

D : Abnormal development with persistence of the stapedial artery.

E: Abnormal development with persistence of a pharyngo-stapedial artery

DA : Dorsal Aorta
VA : Ventral Aorta
I : First Aortic Arch
II : Second Aortic Arch
III : Third Aortic Arch
POA : Primitive Ophtalmic Artery
SA : Stapedial Artery
HA : Hyoïd Artery
CCA : common Carotid Artery
SOA : Supra Orbital Artery
MMA : Middle Meningeal Artery
OB : Orbital Branch
MxMbA : Maxillo-mandibular Artery
VPA : Ventral Pharyngeal Artery
OA : Ophtalmic Artery
PSA : Persistent Stapedial Artery
ICA : internal Carotid Artery
ECA : External Carotid Artery
ITC : Internal tympanic canaliculus
APA : Ascending Pharyngeal artery
ITA : Inferior tympanic artery

Modified from Lasjaunias [1] and Moret [10]

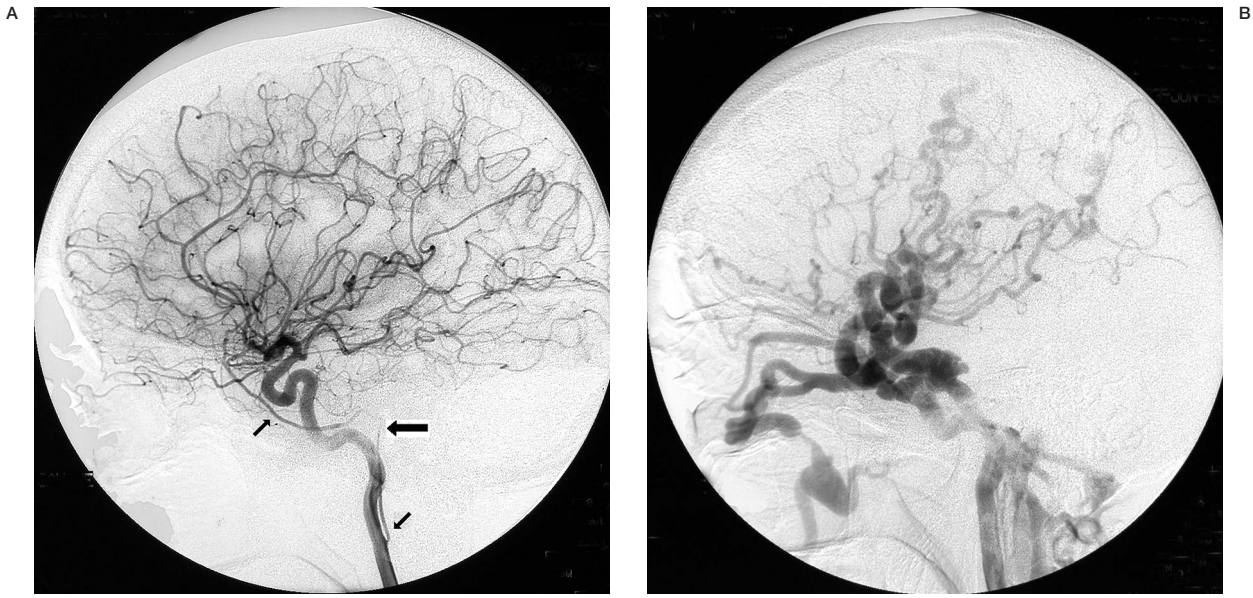

Figure 1 Initial carotid angiogram. A) Left carotid angiogram, lateral view, shows a persistent pharyngo-stapedial artery arising from the cervical internal carotid artery. B) Right carotid angiogram, lateral view, shows a high-flow carotid-cavernous fistula, that drains into the inferior petrosal sinus, superior ophthalmic vein, Brechet sinus, and cortical veins. There is no persistent phayngo-stapedial artery (PSA) visible.

the foramen spinosum and bilateral finding of a small anomalous vessel crossing the middle ear and entering the anterior tympanic segment of the facial nerve canal.

The right carotid-cavernous fistula had obviously to be treated because of the extensive cortical venous drainage, and the endovascular intervention took place four month later. Surprisingly, the pretherapeutic angiogram showed that the fistula had altered in the meantime. A thrombosis of the ophthalmic vein had occurred, leading to a reduction of the drainage and flow through the malformation. The persistence of a pharyngo-stapedial artery also on the right side, suspected on the temporal bone CT imaging, was not evident in the angiogram four month before, because of the vascular "steal" through the arteriovenous malformation. In the new haemodynamic features of the fistula, the homolateral Ph-SA was clearly visible in the angiogram. The patient being under general anesthetic, an envoy 6F introducer catheter was directed into the mid portion of the ICA. The microcatheterism of the cavernous sinus was done through the large fistula with a Prowler Plus and a Terumo 16/90. Complete closure of the fistula was obtained, with preservation of the internal carotid artery using three Guglielmi detachable coils 18, one GDC 10 coil and one matrix 3D coil. Heparin was administered at a curative dose during 72 hours. The patient was discharged five days later with no clinical evidence of carotid-cavernous fistula.

One year after the procedure, follow-up examination showed the persistence of the perfect clinical result. We performed MR angiography with a 3 Tesla General Electric machine that demonstrated that the fistula was still closed. MR angiography also provided us with a precise illustration of the bilateral Ph-SAs, first arising from the cervical ICAs, then following their course in the middle ear, and finally becoming the middle meningeal arteries

Discussion

The stapedial artery is transiently present in normal foetal development, connecting the internal carotid artery to the branches of the future external carotid artery. This small vessel normally regresses in the 24 mm embryo. Its persistence is a rare variant resulting from an error at a precise step during the complex embryologic development of the cervical and cranial vasculature [1].

At an initial stage of development, the ventral aorta and dorsal aorta are anastomosed by

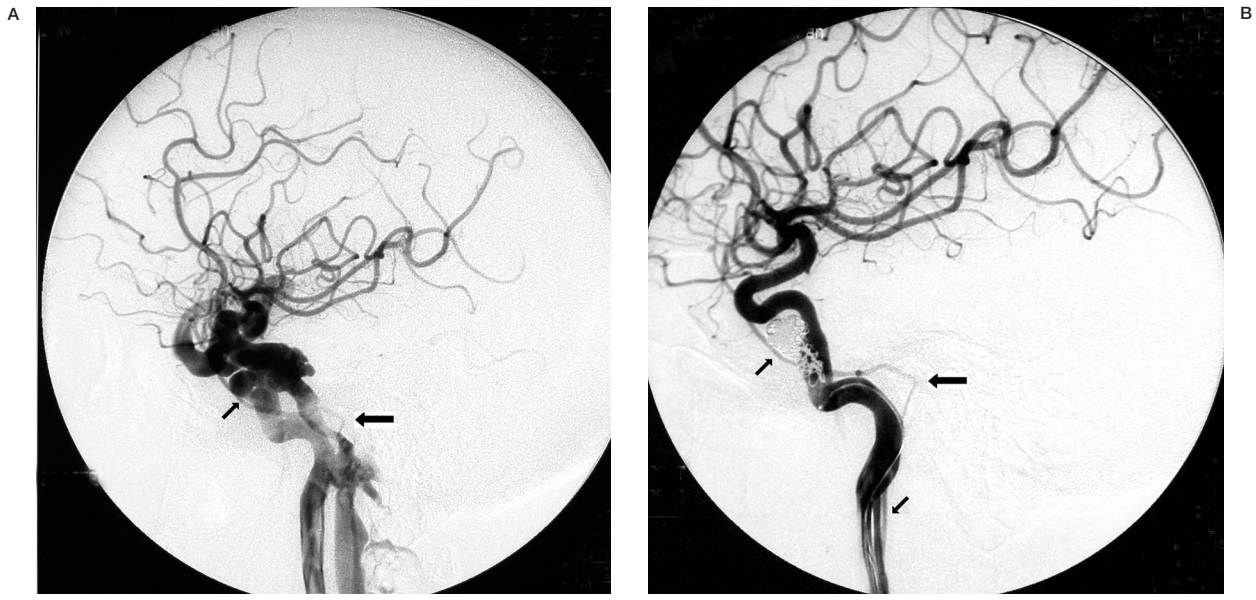

Figure 2 Right internal carotid angiogram performed four months later. A) Pretherapeutic right carotid angiogram, lateral view, shows that the fistula had spontaneously altered in the meantime with occlusion of the ophthalmic vein and reduction of the vascular steal. The homolateral Ph-SA is now opacified (arrows). B) Right carotid angiogram, lateral view, at the end of the procedure. After complete coil occlusion of the fistula with preservation of the ICA the Ph-SA is now better opacified.

a certain number of arterial structures, the aortic arches. Each aortic arch corresponds to a branchial arch. The aortic arches are not all present at the same time, but develop in numerical order in the cranio-caudal direction. The first arch gives rise transiently to the mandibular artery, but later regresses totally. The second arch becomes the hyoid artery, from which arises the stapedial artery in the fourth or fifth week of foetal life. This stapedial artery passes through the future tympanic cavity, pierces the stapes primordium (leaving its imprint as the obturator foramen) and enters the cranial cavity. There, at a later stage of development, it divides into two branches. The upper branch, named supra-orbital artery, remains endocranial and becomes the middle meningeal artery in the normal adult system. The lower branch is the maxillomandibular artery, which leaves the cranial cavity through the foramen spinosum. The third arch gives rise to the ventral pharyngeal artery which represents the future external carotid. Then, the definitive adult system will be set up with a complex atrophic process of two parts of the stapedial artery and with the annexation of its two branches by other vascular structures.

- Intracranially: annexation of the orbital branch of the supra-orbital artery by the primitive ophthalmic artery (an embryonic artery derived from the cranial portion of the dorsal aorta), and regression of the orbital branch above its annexed portion. The persistent second branch of the supra-orbital artery is the future middle meningeal artery.

- Extracranially: annexation by the ventral pharyngeal artery (the future external carotid artery) of the maxillomandibular artery, and simultaneous regression of the tympanic portion of the stapedial artery. Therefore, the flow reverses in the maxillomandibular artery through the foramen spinosum. The arterial flow directed exocranially at first, reverses inward toward the cranial cavity.

The vessels previously supplied by the stapedial artery are then supplied by the external carotid artery and by the ophthalmic artery, and the stapedial artery degenerates during the tenth week. It persists if this complex development fails to occur. The vulnerable point of the system is the portion of the maxillomandibular artery that goes through the foramen spinosum and where the flow normally reverses. If the annexation of this artery by the pharyngeal ascending artery occurs earlier than the regression of the stapedial artery, two opposite flows will be set up in this transcranial branch. This induces blood stagnation and at last regression

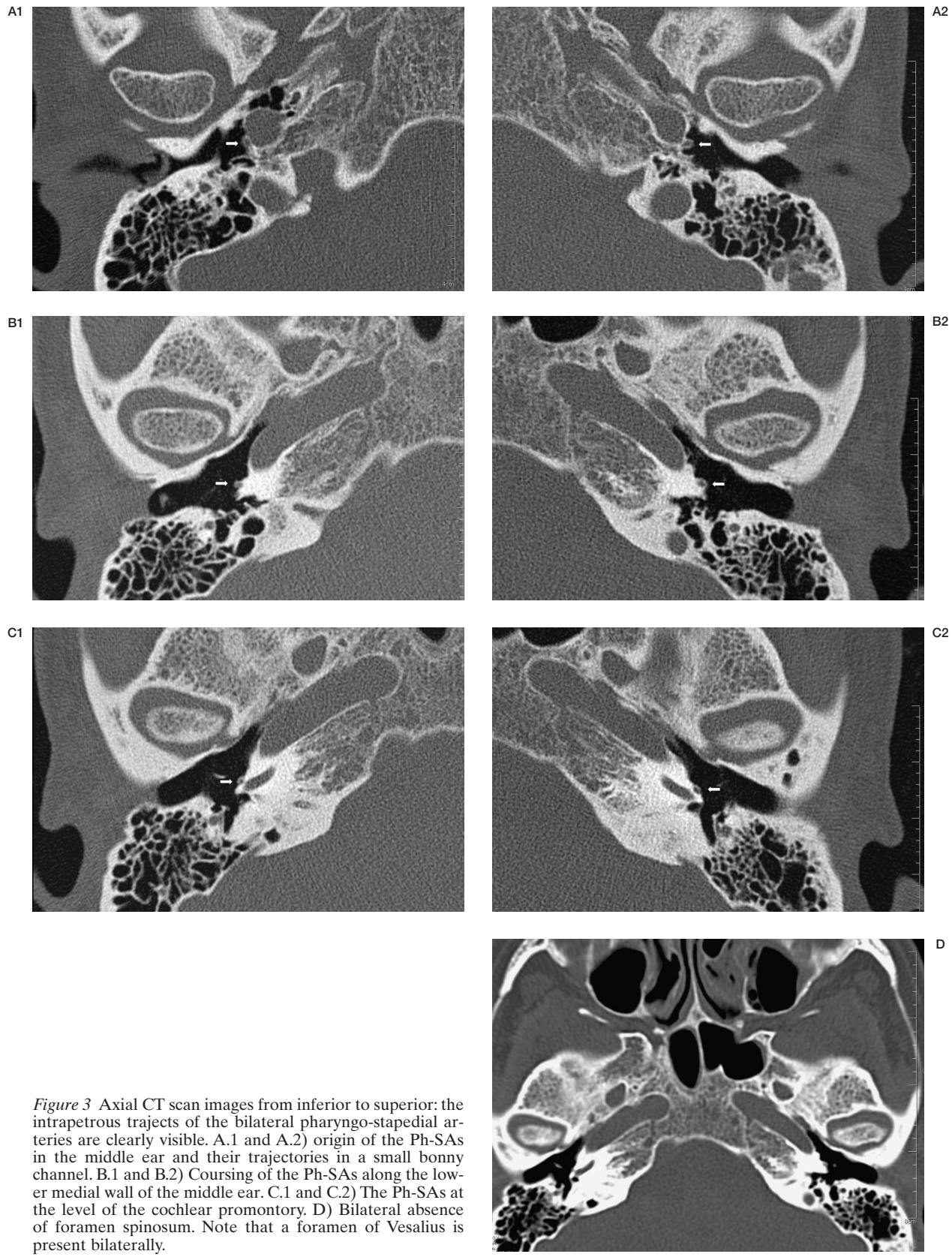

Figure 3 Axial CT scan images from inferior to superior: the intrapetrous trajects of the bilateral pharyngo-stapedial arteries are clearly visible. A.1 and A.2) origin of the Ph-SAs in the middle ear and their trajectories in a small bonny channel. B.1 and B.2) Coursing of the Ph-SAs along the lower medial wall of the middle ear. C.1 and C.2) The Ph-SAs at the level of the cochlear promontory. D) Bilateral absence of foramen spinosum. Note that a foramen of Vesalius is present bilaterally.

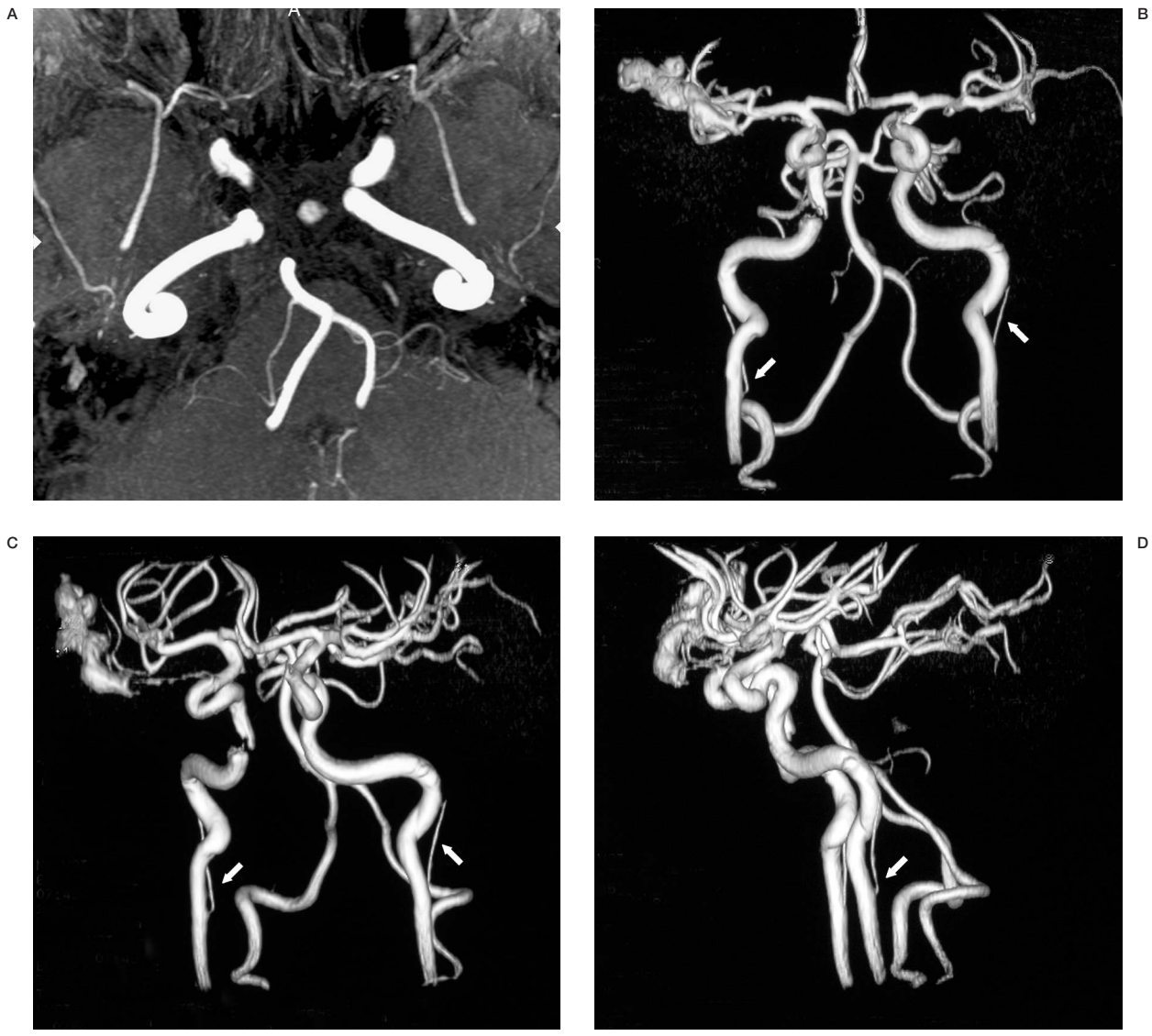

Figure 4 Time-of-flight MR angiogram performed one year after the procedure: 3D and axial MR images show the bilateral persistent pharyngo-stapedial arteries arising from the ICAs and supplying the middle meningeal distribution.

of this transcranial branch and its bony channel, the foramen spinosum[1]. In this case, the tympanic portion of the stapedial artery does not regress and typically supplies the middle meningeal artery. Maxillomandibular branches are supplied by the ECA.

The inferior portion of the PSA arises from the hyoid artery, which does not regress. The pharyngo-stapedial artery is a rare variant that occurs when there is both persistence of the stapedial artery but regression of the hyoid artery at the level of its origin in the ICA. Under these conditions, the blood flow through the stapedial artery is assumed by the inferior tympanic artery – a branch of the ascending pharyngeal artery – via its anastomosis with the distal portion of the hyoid artery along Jacobson's nerve.

A persistent stapedial artery is a 2 mm arterial vessel that arises from the petrous part of the ICA and enters the middle ear cavity by its floor. The pharyngo-stapedial artery arises exocranially from the ascending pharyngeal artery, proceeds upward and enters the tympanic cavity through the inferior tympanic canaliculus, accompanying Jacobson's nerve. Then, in the

middle ear the two variants have the same trajectory. The anomalous vessel goes along a bony ridge to the promontory in a posterior direction; leaving the promontory, it courses through the obturator foramen of the stapes and enter the second portion of the facial canal, briefly accompanying the nerve. The PSA leaves the facial canal just before the geniculate ganglion through a special opening and then travels anteriorly and medially in the extradural space of the middle cranial fossa where it ends by becoming the middle meningeal artery [1,2].

In our observation, an unusual aspect is the direct emergence of the ascending pharyngeal artery from the internal carotid artery. The ascending pharyngeal artery arises more often from the external carotid, but its origin from the ICA has already been described [4].

The first description of a PSA was that of Hyrtl in 1836 [5]. After this post-mortem discovery, a few surgical discoveries were reported [2]. Then, angiographic findings, CT findings [6] and more recently MRA findings were published [7,8]. PSA is a rare congenital vascular anomaly and its prevalence based on stapes surgery series is quoted as one to 4 000 to one to 10 000 (0.02-0.01 %) [2,3]. Most of the cases reported are unilateral. Fourteen previous cases of bilateral PSA have been described [7,9]. It is more often an isolated anomaly but the association with an aberrant ICA is not rare [3,6].

The persistent pharyngo-stapedial artery (or pharyngotympanostapedial artery) has been reported by Lasjaunias and Moret [1,10]. In many studies, this rare variant is probably not distinguished from the "usual" PSA [6]. The pharyngo-stapedial artery and the stapedial artery have in fact the same aspect in the middle ear and the possible pathological consequences are similar.

In our case, the bilateral Ph-SAs were associated with a post-traumatic carotid-cavernous fistula and whereas the controlateral Ph-SA was clearly visible, the homolateral Ph-SA was hidden at first because of the high flow in the fistula. Surprisingly, spontaneous modification of the venous drainage of the fistula arose after laps of time, with closure of the ophthalmic vein. The result of this was the decreasing of the vascular "steal" through the malformation and thus, opacification of the homolateral Ph-SA. Finally, the best visualisation of the homolateral Ph-SA was obtained after complete closure of the fistula.

In the literature, patients reported with persistent stapedial artery are mostly asymptomatic [3]. Otoscopic examination may disclose a reddish pulsatile mass behind the tympanum. When symptomatic, PSA may induce pulsatile tinnitus, or a conductive hearing loss due to stapes ankylosis [11].

Presence of a PSA may complicate any middle ear surgery especially when it is an incidental finding. Its damage during surgery may result in unexpected bleeding. However, some authors have reported several cases of damage to a PSA without post-operative sequelae, leading them to the conclusion that a PSA should not hinder middle ear surgery [9]. Recently, patients suffering from tinnitus associated with a PSA have been successfully treated by ligation, without complication [3,12]. Embolization may also be considered but no experience is reported. Our patient had no clinical evidence of bilateral PSA and we did not propose any treatment.

Conclusions

The persistent stapedial artery and the pharyngo-stapedial artery are rare anatomic variants. These two variants only differ from one another by their origin. Imaging identification of these anomalies is helpful in the evaluation of a pulsatile mass behind the tympanum and of tinnitus and hearing loss. Absence of the foramen spinosum is an indirect sign of PSA or Ph-SA, but it is seen in up to 3% of skull base CT studies, since it can be a normal variation, particularly when the middle meningeal artery arises from the ophthalmic artery [13]. Direct identification of PSA can be made by the use of high resolution temporal bone CT and conventional angiography. A couple of recent studies have reported MR angiographic findings of PSA [7,8]. We report the easy detectability of a stapedial artery on 3 Tesla MR angiograms.

It is useful to predict the anomalous vessel prior to tympanotomy, but recent studies conclude that injury to this artery or even complete section probably would not cause major sequelae; Patients suffering from tinnitus and hearing loss have been successfully treated with ligation of a PSA. Endovascular techniques should also be considered.

References

1 Lasjaunias P, Berenstein A: Surgical Neuro-angiography. Springer-Verlag, Berlin Heidelberg 1987.
2 Guinto FC, Garrabrant EC, Radcliffe WB: Radiology of the Persistent Stapedial Artery. Radiology 105: 365-369, 1972.
3 Silbergleit R, Quint DJ, Mehta BA et Al: The Persistent Stapedial Artery. Am J Neuroradiol 21 : 572-577, 2000.
4 Lefournier V, Vasdev A, Bessou P et Al: A Persistent Pharyngohyostapedial artery : Embryologic implications. Am J Neuroradiol 20: 271-274, 1999.
5 Hyrtl J: Neue Beobachtungen aus dem Gebiete der menschlichen und vergleichenden Anatomie. Med Jahrb d Oesterreich Staates 10: 457-466, 1836.
6 Thiers FA, Sakai O et Al: Persistent Stapedial Artery: CT Findings. Am J Neuroradiol 21: 1551-1554, 2000.
7 Roll JD, Urban MA et Al: Bilateral Aberrant internal Carotid Arteries with Bilateral Persisent Stapedial Arteries and Bilateral Duplicated Internal Carotid Arteries. Am J Neuroradiol 24: 762-765, 2003.
8 Yilmaz T, Bilgen C, Savas R: Persistent Stapedial Artery: MR Angiographic and CT Findings. Am J Neuroradiol 24: 1133-1135, 2003.
9 Govaerts PJ, Marquet TF, Cremers WR: Persistent Stapedial Artery : does it prevent successful surgery? Ann Otol Rhinol Laryngol 102: 724-728, 1993.
10 Moret J, Delvert JC, Lasjaunias P: "Abnormal" vessels in the middle ear. J Neuroradiology 9: 227-236, 1982.
11 Jain R, Gandhi D, Gujar S et Al: Case 67: Persistent stapedial artery. Radiology 230: 413-416, 2004.
12 Murphy MP, Seidman MD, Baugh RF: Can a Persistent Stapedial Artery be Safely and Effectivly Removed? A Case Report with Therapeutic Implications. Int Tinnitus J 1: 147-152, 1995.
13 Ginsberg LA, Pruett SW, Chen MY et Al: Skull base foramina of the middle cranial fossa: reassessment of normal variation with high resolution CT. Am J Neuroradiol 15: 283-291, 1994.

J. Jehl, M.D.
Department of Neuroradiology
Department of Orthopaedic Surgery
University Hospital Jean Minjoz,
Besançon, France